

**In the Claims:**

This listing of claims will replace all prior versions, and listings, of claims in the application:

1-18 (Cancelled)

19. (Currently Amended) A method of determining the presence of a Mucopolidosis IV **deletion mutation mutant** sequence in a nucleic acid, comprising,

a) contacting the nucleic acid with[[;]] :

i) a first oligonucleotide primer that comprises a sequence complementary to a 15-30 bp segment of DNA between positions 100-500 of the MCOLN1 gene (**SEQ ID NO: 8**),

ii) a second oligonucleotide primer that comprises a sequence complementary to a 15-30 bp segment of DNA between positions 6956-7356 of the MCOLN1 gene (**SEQ ID NO: 8**), **and**

iii) an oligonucleotide probe that comprises a sequence complementary to a 13-30 bp segment of a fragment that is amplified using the first and second primer, wherein said probe is labeled with a detectable label which comprises a donor fluorophore and a quencher moiety, wherein said quencher moiety is optionally an acceptor fluorophore; **and**

b) conducting amplification by temperature cycling and monitoring the accumulation of amplified nucleic in real time by detecting an increase in donor fluorophore fluorescence or a decrease in acceptor fluorophore fluorescence which indicates the presence of the Mucopolidosis IV mutant sequence in the nucleic acid.

20. (Original) The method of claim 19 wherein the first oligonucleotide primer comprises a sequence that consists essentially of 5'-CTT GCT CTG TTG CCC AGG CT -3' (SEQ ID NO. 3).

21. (Currently amended) The method of claim 19 wherein the second oligonucleotide primer comprises a sequence that consists essentially of **[[and]]** 5'-CTC ACC GTG CTG GAA GAC ACT -3' (SEQ ID NO. 4).

22. (Currently amended) The method of claim 19 wherein the probe comprises a sequence that consists essentially of ~~and the probe in ii)~~ **is** 5'- AGACC CAG GCC CAC AT- 3' (SEQ ID NO: 7).

23. (Original) The method of claim 19 wherein the donor fluorophore is 2'-chloro-7'-phenyl-1,4-dichloro-6-carboxyfluorescein (VIC), 6-carboxyfluorescein (FAM) or tetrachloro-6-carboxyfluorescein (TET).

24. (Original) The method of claim 19 wherein amplification by temperature cycling is with a DNA polymerase with 5' exonuclease activity and wherein binding of the probe to amplified nucleic acid results in degradation of the probe during DNA synthesis.

25. (Currently amended) A method of detecting the presence of one or two Mucopolipidosis IV mutant sequences in a nucleic acid, comprising,

a) contacting the nucleic acid with**[[;]]** :

i) a first oligonucleotide primer that comprises a sequence complementary to a 15-30 bp segment of DNA between positions ~~100-500~~ **5124-5524** of the MCOLN1 gene **(SEQ ID NO: 8)**,

ii) a second oligonucleotide primer that comprises a sequence complementary to a 15-30 bp segment of DNA between positions ~~6956-7356~~ **5541-5941** of the MCOLN1 gene **(SEQ ID NO: 8)**,

iii) a first oligonucleotide probe that comprises a sequence complementary to a 13-30 bp segment of a fragment that is amplified using the first and second oligonucleotide primers, wherein said **first oligonucleotide** probe **includes position 5534 of the MCOLN1 gene** **(SEQ ID NO: 8)**, **wherein said first oligonucleotide probe** is labeled with a first detectable

label which comprises a donor fluorophore and a quencher moiety, wherein said quencher moiety is optionally an acceptor fluorophore[[:]] :

iv) a third oligonucleotide primer that comprises a sequence complementary to a 15-30 bp segment of DNA between positions 100-500 of the MCOLN1 gene **(SEQ ID NO: 8)**,

v) a fourth oligonucleotide primer that comprises a sequence complementary to a 15-30 bp segment of DNA between positions 6956-7356 of the MCOLN1 gene **(SEQ ID NO: 8)**, and

vi) a second oligonucleotide probe that comprises a sequence complementary to a 13-30 bp segment of a fragment that is amplified using the third and fourth primers, wherein said probe wherein said **second oligonucleotide** probe is labeled with a second detectable label which comprises a donor fluorophore and a quencher moiety, wherein said quencher moiety is optionally an acceptor fluorophore, and wherein said second detectable label is distinguishable from said first detectable label; **and**

b) conducting amplification by temperature cycling and monitoring the accumulation of amplified nucleic in real time by detecting an increase in donor fluorophore fluorescence or an increase or decrease in acceptor fluorophore fluorescence, which indicates the presence of one or both of the Mucopolipidosis IV mutant sequences in the nucleic acid, **wherein said first and second primer and first probe detect a single base transition mutation and said third and fourth primer and second probe detect a deletion mutation.**

26. (Original) The method of claim 25 wherein the first oligonucleotide primer comprises a sequence that consists essentially of 5'-AGC GGG CCG GAC TCA-3' (SEQ ID NO. 1).

27. (Original) The method of claim 25 wherein the second oligonucleotide primer comprises a sequence that consists essentially of 5'-TAA CCA CCA TCG GAT CAA TGT C-3' (SEQ ID NO. 2).

28. (Original) The method of claim 25 wherein the first probe comprises a sequence that consists essentially of 5'-CTGC CCA CGG TAC CT -3' (SEQ ID NO: 6).

29. (Original) The method of claim 25 wherein the third oligonucleotide primer comprises a sequence that consists essentially of 5'-CTT GCT CTG TTG CCC AGG CT -3' (SEQ ID NO. 3).

30. (Original) The method of claim 25 wherein the fourth oligonucleotide primer comprises a sequence that consists essentially of 5'-CTC ACC GTG CTG GAA GAC ACT -3' (SEQ ID NO. 4).

31. (Original) The method of claim 25 wherein the second probe comprises a sequence that consists essentially of 5'-AGACC CAG GCC CAC AT- 3' (SEQ ID NO: 7).

32. (Original) The method of claim 25 wherein the first or second donor fluorophore is 2'-chloro-7'-phenyl-1,4-dichloro-6-carboxyfluorescein (VIC), 6-carboxyfluorescein (FAM) or tetrachloro-6-carboxyfluorescein (TET).

33. (Original) The method of claim 25 wherein amplification by temperature cycling is with a DNA polymerase with 5' exonuclease activity and wherein binding of the probe to amplified nucleic acid results in degradation of the probe during DNA synthesis

34. (Original) The method of claim 25 wherein said nucleic acid containing sample is also contacted with a third oligonucleotide probe comprising a sequence consisting essentially of 5'-TCTG CCC ACA GTA CCT -3' (SEQ ID NO: 5) that hybridizes to a wildtype sequence, wherein said third probe is labeled with a detectable label comprising a donor fluorophore and a quencher moiety wherein said quencher moiety is optionally an acceptor fluorophore, and wherein said third detectable label is distinguishable from said first and second detectable labels.

35-36. (Cancelled)